Refer to: Falk PM: Constrictive pericarditis presenting as a mediastinal mass. Calif Med 119:83-85, Aug 1973

Constrictive Pericarditis Presenting as a Mediastinal Mass

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CHRONIC CONSTRICTIVE PERICARDITIS is characterized by pronounced venous distension with severe dyspnea, ascites out of proportion to the dependent edema, muffled heart tones and usually the absence of significant cardiomegaly on x-ray films. As emphasized by Plauth et al[1] a few patients may present with generalized edema and other signs of hypoproteinemia as a result of a protein-losing enteropathy secondary to the pericarditis. Attention has not been drawn to the fact that the radiologic findings consistent with this diagnosis may be substantially altered by associated defects of the pericardium.

The purpose of this report is to describe a 16-year-old boy with chronic constrictive pericarditis who also had a partial pericardial defect. The resultant herniation of the left atrial appendage caused radiologic manifestations highly suggestive of a mediastinal mass lesion.

Report of a Case

A 16-year-old Mexican boy was admitted to the UCLA Medical Center with complaint of "chest pains and chest pounding" of eight months' duration. The chest pain was sharp in character. Five months before admission here, the symptoms had become more severe and were aggravated by physical activity. There was no history of congenital heart disease, heart murmur, cyanosis, rheumatic fever or rheumatic heart disease. The patient was then taken to an orthopedic clinic in Calexico, where he was being followed for paraplegia secondary to poliomyelitis contracted at three years of age. Chest roentgenogram demonstrated a left hilar mass. Tuberculosis was apparently excluded and it was then that he was referred to the UCLA Medical Center for further study.

On physical examination he appeared thin, short and chronically ill, with a protuberant abdomen and mild respiratory distress. He was mildly febrile (37.7°C) with a regular heart rate of 104 per minute, a respiratory rate of 28 per minute, and blood pressure of 90/64 mm of mercury. The blood pressure did not vary abnormally with respiration. Positive physical findings included facial, sacral and lumbar edema, pink nail beds with clubbing of the fingers, and distended veins over the thorax and abdomen. The neck veins were distended and pulsated vigorously. The lungs were clear to percussion and auscultation. No murmurs were heard. The heart tones were somewhat muffled and an intermittent third sound was heard which varied with respiration (this was later believed to be a "pericardial knock"). The abdomen was distended and there was a positive fluid wave. The liver was palpable 3 cm below the right costal margin. Neurological examination was unremarkable except for generalized muscle wasting with paralysis and absence of deep tendon reflexes in the lower extremities.

Laboratory data included hemoglobin of 12.4 grams per 100 ml of blood, a hematocrit of 37.1 percent, a leukocyte count of 6,100 per cu mm, with 81 segmented neutrophils, 2 banded neutrophils, 8 eosinophils, 1 basophil, 7 lymphocytes, and 1 mononuclear cell. Urinalysis was negative and creatinine content was 0.5 mg per 100 ml. The total serum protein was 3.7 grams per 100 ml with albumin of 2.7 grams. Skin tests for coccidioidomycosis, blastomycosis, histoplasmosis, and tuberculosis (with intermediate and second strength purified protein derivative) were negative. A roentgenogram of the chest showed a mass-like lesion in the mediastinum (Figure 1). No abnormality was noted on aspiration of bone marrow. A cardiac scan was consistent with pericardial effusion but a solid mediastinal tumor could not be ruled out. A roentgenographic bone survey showed no metastatic lesions. A barium swallow with fluoroscopy showed the mass to be a pulsating structure which beat paradoxically with the left ventricle. Electrocardiography revealed gen-

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Submitted October 6, 1972.

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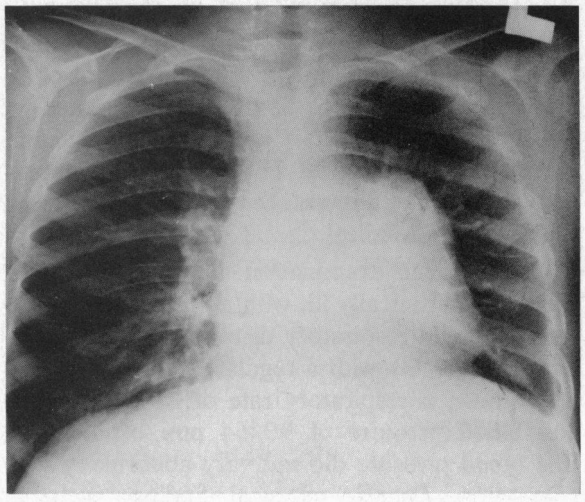

Figure 1.—Teleroentogram showing some cardiomegaly with irregular enlargement of left border suggestive of a mediastinal mass.

eralized low voltage with a prolonged P-R interval. There was evidence of left atrial enlargement (Figure 2).

Cardiac catheterization and angiocardiography demonstrated a large left atrium and aneurysmal dilatation of the left atrial appendage, which appeared to have herniated through a pericardial opening. The antecubital venous pressure was 23 cm of water.

A tentative diagnosis of constrictive pericarditis with partial pericardial defect was made, but some observers felt that a mass lesion was still a possibility. Exploratory operation was recommended and was carried out 17 days after admission. At thoracotomy, thick fibrinous adhesions were found in the left hemithorax. The right hemithorax was free of involvement. There was a tight fibrinous exudate and an adhesive layer between the epicardium and the pericardium. There was herniation of the left atrial appendage through a partial pericardial defect. Radical pericardiectomy was performed. Granulomatous inflammation with calcification and fibrosis of the pericardium was seen on pathologic examination of the surgical specimen. The cause was not established. Postoperatively, congestive heart failure developed and was treated with diuretics, salt-poor albumin, and digoxin. The course thereafter was uneventful. At a later date, acid-fast bacteria were grown from a urine specimen. Also, reexamination of the pericardial pathologic sections revealed the presence of acid-fast bacteria. The patient was discharged with antituberculosis chemotherapy.

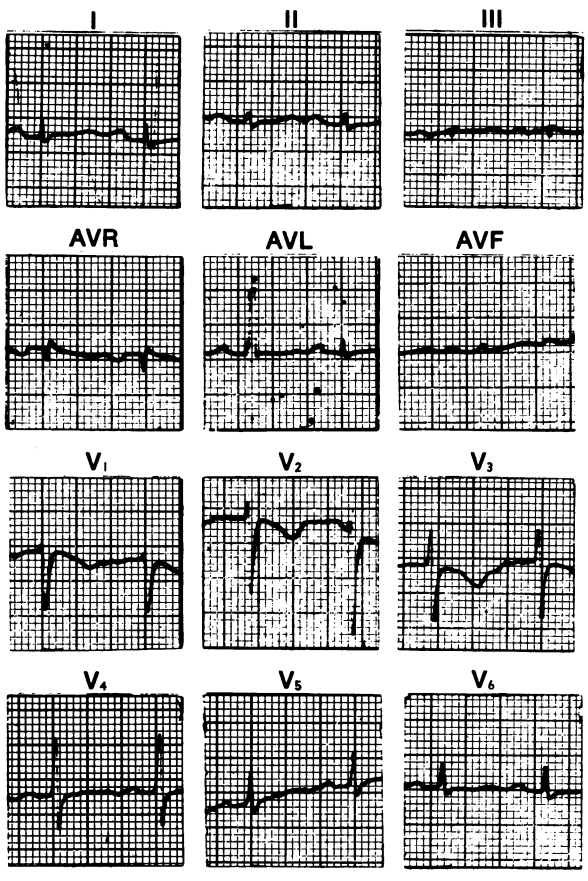

Figure 2.—Electrocardiogram showing decreased voltage in all leads. There is also prolongation of the P-R interval with widening of the P waves suggestive of left atrial enlargement.

Discussion

Constrictive pericarditis is an uncommon pathologic condition in children. According to Jones et al[2] fewer than a hundred cases have been reported in children. The great majority (60 percent) are idiopathic but a significant number (30 percent) are due to tuberculosis.[3]

Pericardial defects are more often found on the left side. They may be partial or total and are congenital in origin. Generally asymptomatic, pericardial defects may be commoner than they are generally believed to be. Abbott's classic review of 1,000 autopsies lists 30 cases of this defect. If symptomatic, they may be associated with vague chest pain and, in rare instances, sudden death possibly due to cardiac herniation and strangulation.[4,5]

The combination of these two conditions has been reported in only one case, that of a 67-year-old woman who was treated successfully by operation.[6] Prominence of the left heart border was ob-

served on a roentgenogram of the chest and was interpreted as indicating left atrial enlargement or dilation of the pulmonary artery segment. Associated constrictive pericarditis was suspected before operation.

The specific cause of the pericarditis in the patient herein reported upon was finally established as tuberculosis. Greenberg et al[6] suggested that the mechanism in the case they reported (non-tuberculous) may have been (1) inflamation of the pericardium resulting from spread of infection from the pleural cavity and (2) adhesions arising from impaired pericardial cushioning during the cardiac cycle.

Chronic constrictive pericarditis is a fatal disease if untreated. It can be corrected surgically, however, and recognition of it is therefore important. When it is associated with a pericardial defect, the radiologic features may be misleading. Although only one previous instance of such an association could be found in the literature, this may not reflect the true incidence, as congenital pericardial defects may be commoner than is generally believed.

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